Modern Concepts of Cardiovascular Disease

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THE CLINICAL ASPECTS OF CONGENITAL CARDIAC DISEASE

Part II

SPECIAL FEATURES

PATHOGENESIS OF THE DEFECT

As indicated in Part I, the chief proximal cause of congenital heart disease is: (a) *Arrest of Development*, and the earlier in embryonic life this occurs, the graver and more complex the anomaly. Most cases of so-called *morbus coeruleus* (Group III) belong in this category, in that interruption has occurred between the fifth and eighth week of embryonic life, at that highly critical period before the cardiovascular septa have come into apposition, or the bulbus cordis has undergone involution, or torsion of the great trunks is complete. Interference with normal growth at this time commonly leads to a series of anatomical adjustments resulting in multiple associated anomalies. This explains those curious combinations of complete or partial ventricular septal defect with dextroposition or transposition of the aorta, regular division of the great trunks, suppression of parts and those confusing examples of incomplete heterotaxy of chambers which reveal themselves in the light of comparative embryology as simple and beautiful structural adaptations to the needs of the altered circulation. Arrest at a slightly later stage after the complex changes at the base of the heart have been consummated, but *before* the cardiovascular septa have closed (end of eighth week) results in the localized septal defects of the *cyanose tardive* (Group II), while arrest *after* their closure causes only those minor anomalies leading to cardiovascular strain (Group I).

(b) Foetal disease plays an apparently minor role in the etiology. An early myocarditis may, however, be the underlying factor in some cases of very early embryonic arrest. In others, the ultimate cause may lie in the maternal environment of the developing embryo; or in an inherent weakness of the germ plasm due to alcoholic toxins, consanguinity, etc.; or in some still more obscure hereditary factor.

CYANOSIS

The familiar symptom-complex of congenital cyanosis develops when oxygen-unsaturation is permanently raised above the threshold value at which this becomes manifest in the capillaries (6.7 volumes per cent). It consists in a progressively increasing bluish discoloration of the skin and mucous membranes, clubbing of the nose and extremities, capillary hyperplasia, cyanosis retinae, dyspnoea, dysphoeic attacks and polycythemia. Calculation of the venous shunt which exists in these cases can be done by the Van Slyke technique, as explained in Lundsgaard's and the writer's Monographs.

THE DIFFERENT DEFECTS

The following table shows the relative frequency in the writer's statistics of some of the more important cardiac anomalies:

I. *Acyanotic Group. No abnormal communication (mostly left-sided lesions).* Pericardial defect 44; so-called idiopathic hypertrophy 17; subaortic stenosis 12; aortic or mitral stenoses or atresias 72; bicuspid aortic valve 78; coarctation, adult type 105; right or double aortic arch 40; right subclavian from descending aorta 7; left coronary from pulmonary artery 10.

II. *Cyanose Tardive Group. Cases of arterial-venous shunt with transient or terminal reversal of flow.* Defects of auricular septum 81; patent foramen ovale 290. Defects of interventricular septum 264. Defects of aortic septum 22. Patent ductus arteriosus 242.

III. *Cyanotic Group. Cases of permanent venous-arterial shunt or retardation of flow.* Pulmonary stenosis with closed septum 31; with septal defect 120; pulmonary

astresia 59; transposition of great trunks 111; cortriloculare 56; persistent truncus 14; incomplete heterotaxy 5.

DIAGNOSIS

This problem must be approached from a threefold aspect. *First*, a *congenital* is to be distinguished from an *acquired* lesion. Significant of the former are: The general habitus of the patient and the frequent incongruous appearance of X-ray and electrocardiogram; the history dating from infancy and the absence of etiological factors; the presence of atypical physical signs, differing in coarse, rough character, high-pitched timbre, seat and zone of transmission from those of postnatal valvular disease; and the familiar symptomatology of cyanosis when raised oxygen-unsaturation results from the defect. *Secondly*, the case should be assigned to its place in the Clinical Classification. In Group I, the *absence* of cyanosis and the fact that these are mostly "left-sided lesions" as revealed by X-ray and electrocardiogram are sufficiently differential points. In Group II, the *absence of cyanosis* in older subjects, except as a terminal or transient phenomenon, and in Group III, the *persistent presence of cyanosis* with its attendant symptomatology, are distinctive; and here the electrocardiogram is again useful as revealing the extreme right preponderance of the "right-sided lesions" in almost all cases of true morbus coeruleus. It must be remembered, however, that even in grave cases cyanosis frequently does not manifest itself for some months after birth, so that in children under two years its absence cannot be taken as a sure criterion of the nature of the defect. *Thirdly*, the diagnosis must be attempted of the individual defect. The following may present distinctive features:

Group I. In *coarctation of the aorta* signs of hypertension in the upper part of the body with reduced blood pressure in the lower extremities, tortuous dilated pulsating collaterals over the thorax and erosion of the ribs as seen by the X-rays are pathognomonic; so also in *subaortic stenosis* are the harsh systolic murmur and coarse thrill localized at the second right interspace in the absence of constitutional symptoms of aortic obstruction; and in *right aortic arch* the broad aortic shadow extending to the right clavicle with esophagus curving to the left seen under barium X-ray technique.

Group II. All the forms of cardiovascular defects can usually be diagnosed: In localized *defect at the base of the interventricular septum* the prolonged rather rough or high-pitched holosystolic murmur with accompanying thrill with occasionally partial heart block, and the entire absence of cyanosis in a healthy young adult are usually pathognomonic; so also in *patent ductus arteriosus* is a machinery murmur and thrill over the left base of the heart with X-ray cap and other signs of pulmonary dilatation, hypoplasia of the aorta, some cardiac enlargement, and in some cases, transient cyanotic attacks. The same clinical picture applies to the rarer cases of *aortic septal defect*, but here the roaring character of the machinery murmur in the third left interspace with frequently diastolic accentuation from pulmonary insufficiency, are significant of the position of the defect between the root of the aorta and base of the pulmonary artery or right ventricle close to the chest wall. Most intriguing of all from the diagnostic standpoint are the *defects of the auricular septum and patent foramen ovale*, in which the habitus of the patient indicating hypoplasia of the aorta, the great pulmonary arterial dilatation and enlargement of the right auricle as shown in the high P wave, together with a history of pulmonary infarction and absence of cyanosis except as a terminal phenomenon are characteristic, especially when accompanied by a distant, sometim inconstant, presystolic-systolic murmur over t third and fourth interspaces with sometimes a fa thrill.

Group III. The *bizarre* combination of defec that may underlie the pronounced symptomatolo makes differential diagnosis problematical, exce in certain classic cases of pulmonary stenosis. the tetralogy of Fallot (pulmonary hypoplasia wi septal defect and dextroposition of the aorta), whi is the commonest cause of congenital cyanosis young adults, the rough systolic murmur and coar thrill at the second left interspace characteristi of this lesion are usually less pronounced than i cases with closed septum, and the thrill may ev be absent while the murmur may be transmitte into the neck or heard in the back. Clubbing is in tense, and X-ray shows widening at the right ba with typical *coeur-en-sabot* form at the left. Th apex curves upward owing to the great hypertrop of the right ventricle and pulmonary conus. This shown also by the marked right preponderance an high P wave.

COMPLICATIONS AND SEQUELAE

The greatest danger that threatens congenita cardiac patients who survive childhood is the deve opment of *an infective process in the vicinity of th defect.* This contingency menaces especially th subjects of *localized aortic* or *intraventricular septa defect* or *patent ductus*, in whom the continuou arterial-venous shunt induces a fibrosis about it margins especially on the right side and on th adjacent and opposite wall of the right ventricle an pulmonary artery, thus providing a seat of electio for the localization of an infective process. The sam applies to *congenitally bicuspid aortic valve*, whic inevitably undergoes sclerosis, and supplies an ide nidus for micro-organisms which almost invariabl becomes the seat of subacute bacterial endocarditi These patients, therefore, who have otherwise a good expectation of life, are to be regarded a "potential cardiacs," in the same sense as are th subjects of "healed" rheumatic lesions.

Auricular septal defects escape this danger, bein practically never the seat of an acute inflammator process. A rare complication that may occur i these cases is *paradoxical embolism.* Both this acci dent and *cerebral abscess* may occur also in septa defect with pulmonary stenosis (tetralogy of Fa lot). *Spontaneous rupture of the aorta* terminate 17 per cent of 212 cases of coarctation analyzed; another 11 per cent of the same series death too place by *cerebral hemorrhage.* In the cyanotic grou *sudden death* without apparent cause is relativel frequent (as also in coarctation) or *pulmonary tu berculous* may be the terminal factor. But th commonest of all causes in these casts is failin compensation.

PROGNOSIS AND TREATMENT

The prognosis in Groups I and II is good a regards the lesion itself, which has little bearin upon the duration except for its tendency to indu the complications, which hang like a drawn swor over these subjects. The treatment, therefore, i largely preventive as in other potential cardiac In Group III on the other hand, the course canno but be progressively downward, for the permanent raised oxygen-unsaturation creates a vicious circ in which thickened pulmonary capillaries, tox metabolites and polycythemia act and react wit oxygen-want to increase the discomfort of thes unfortunate subjects. Here the treatment can onl be palliative and directed to avoidance of com plications.

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